Case Study Presentation: Craniopharyngioma

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History

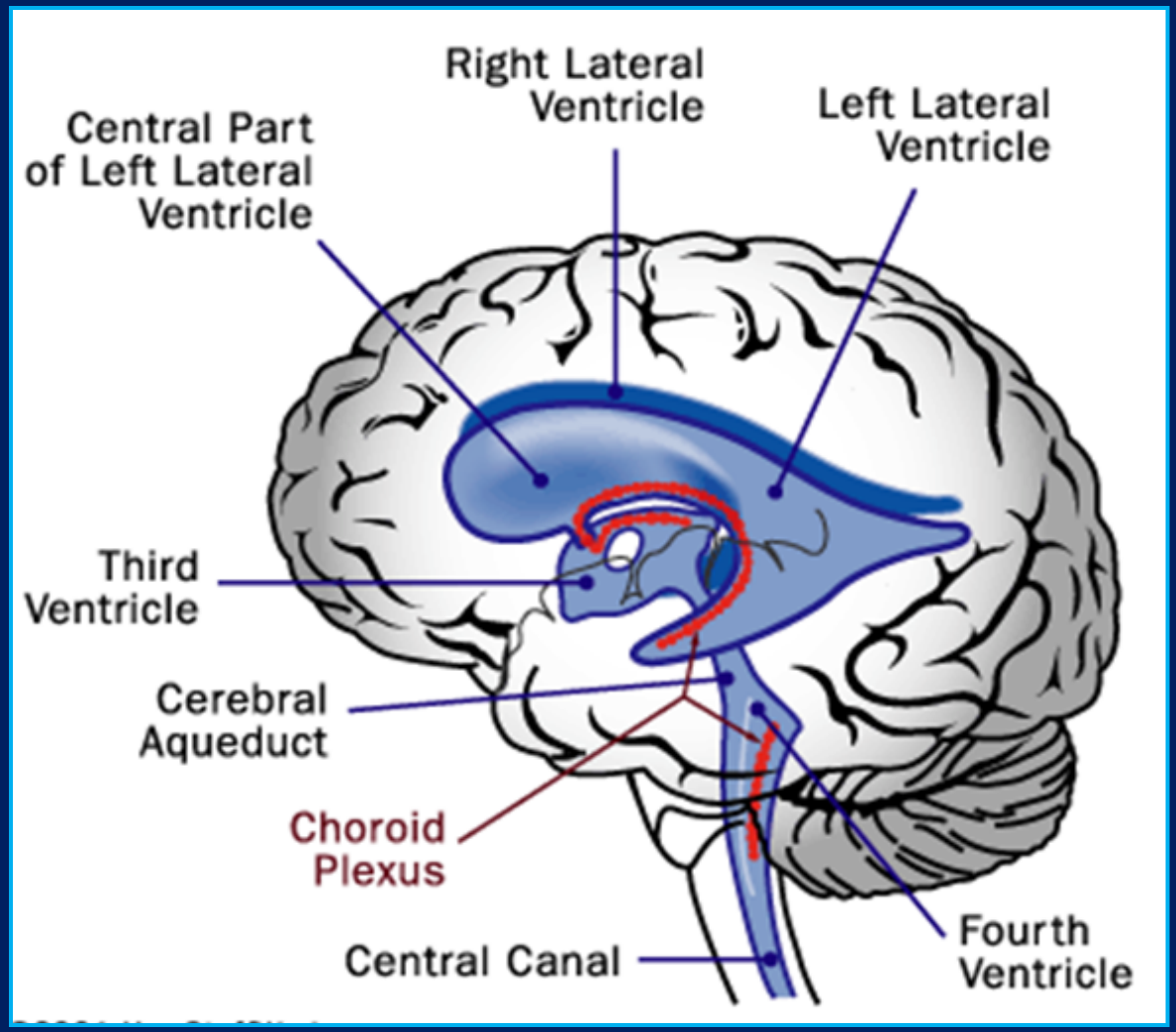
- 17 year old boy - Luke
- 4 year history of fatigue
  - Had swine flu age 13yrs, and ‘never regained energy’
  - Complained of headaches, but ‘ignored as family history of migraines’
- Presented with a 6 week history of peripheral visual field loss – left, then right

Brain MRI - Craniopharyngioma

Brain MRI results

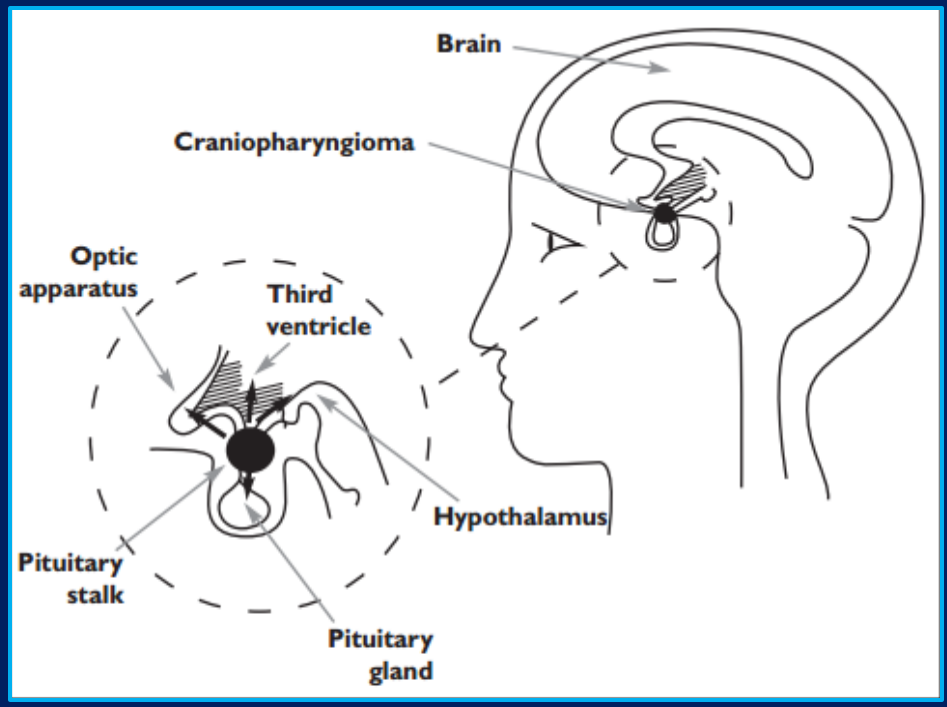
- Mostly cystic post-chiasmatic suprasellar mass, with a small solid calcified component anteriorly and on the right side.
- It elevates the anterior visual pathways and slightly favours the right side without eroding through the floor of the third ventricle
- Appearances are typical of a craniopharyngioma

The Ventricular System of the Brain



Craniopharyngioma

- Benign brain tumour
- Thought to form and grow from some misplaced cells in the brain close to the pituitary gland
- Presentation varies
  - Most commonly occurring symptoms are headaches and blurred vision

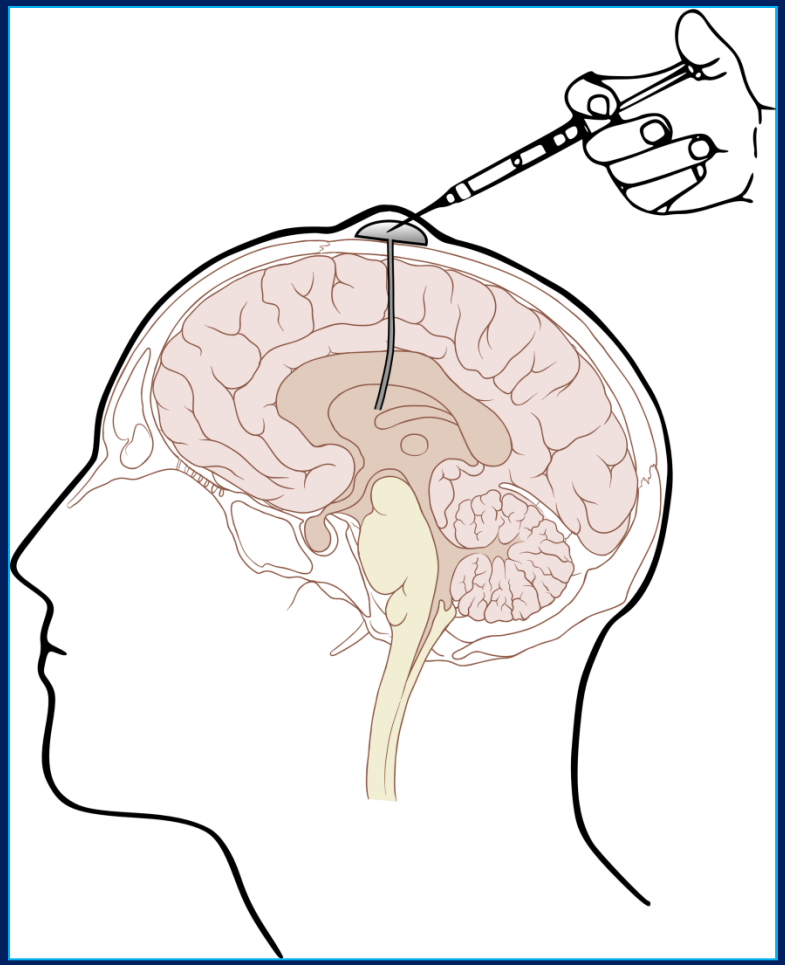


Surgery

- 5 March 2014
  - Cyst aspiration
    - 12mls of fluid
  - Right sided ommaya reservoir inserted
  - Bi-temporal hemianopia recovered following surgery within 24 hours

Ommaya reservoir

- Intraventricular catheter system
  - Can be used for aspiration of cerebro spinal fluid (CSF), or delivery of drugs
  - Indwelling catheter into one of the lateral ventricles in the brain



Post op endocrine assessment

- Insulin Tolerance Test 14 March 2014
- Intact pituitary reserve except severe growth hormone deficiency
- Good peak cortisol
- Normal thyroid function
- Normal gonadal function
- No diabetes insipidus

Insulin Tolerance Test - results

T =	- 0h20m	0h00m	0h20m	0h30m Glucogel	60m	90m	1h20m
Glucose (mmol/L)	5.2	5.0	3.4	2.5	6.3	8.4	7.8
GH (mcg/L)	<0.05	0.06		0.88	1.54		0.08
Cortisol (nmol/L)	377	469		307	319	226	162

- Growth hormone therapy commenced April 2014
- Nordipen 1.0mg daily s/c

Neurosurgical follow up

- Post operative MRI scan
  - Small solid component at the anterior wall of the cyst
  - Intimately associated with the pituitary stalk, the carotid vessels and the optic chiasm
- Complete surgical excision would be difficult
  - Either transphenoidal or transcranial
- Radiotherapy proposed
  - April 2014 – Proton beam therapy, Jacksonville, Florida
    - Tom too old under NHS guidance as over 16
      - Family planned to raise funds for treatment in USA
      - Eligible for clinical trial
        - » Month of pre treatment tests = treatment paid for

Further post operative follow up

- 27 March 14
  - Sperm cryopreservation
- 6 May 2014 - Florida
  - Standard Short Synacthen Test
    - Cortisol: 0840 180nmol/l → 0900 380nmol/l
    - FT4 0.7ng/dl, TSH 0.7mIU/ml
- Commenced:
  - Hydrocortisone (oral) 5mg three times a day
  - Thyroxine (oral) 75mcg once a day

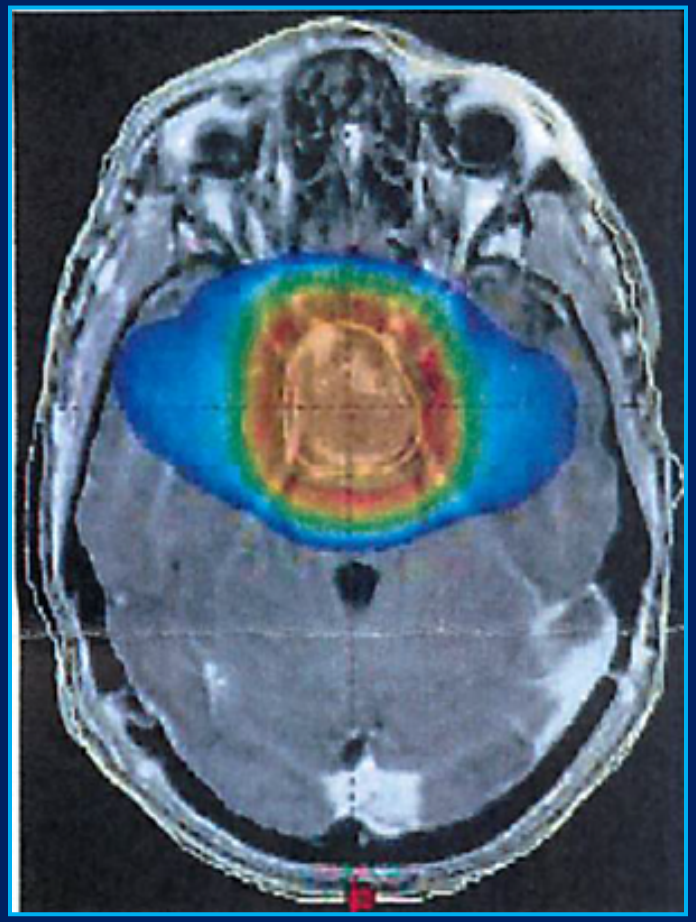
Proton beam treatment

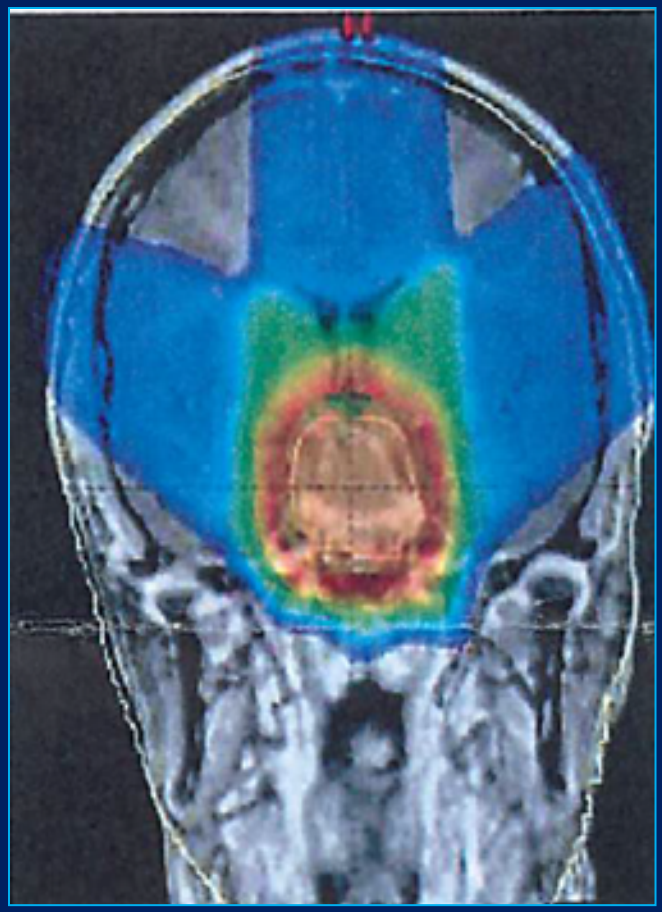
- Type of external beam radiotherapy that uses ionizing radiation
- Tumour is targeted with a beam of protons
- Protons
  - Damage the DNA of the cells, halting the reproduction
  - Due to their large mass
    - Little lateral side scatter in the tissue
    - All have a certain range
      - Very few go beyond that distance
- ? Overall benefit over conventional radiotherapy

Proton beam treatment

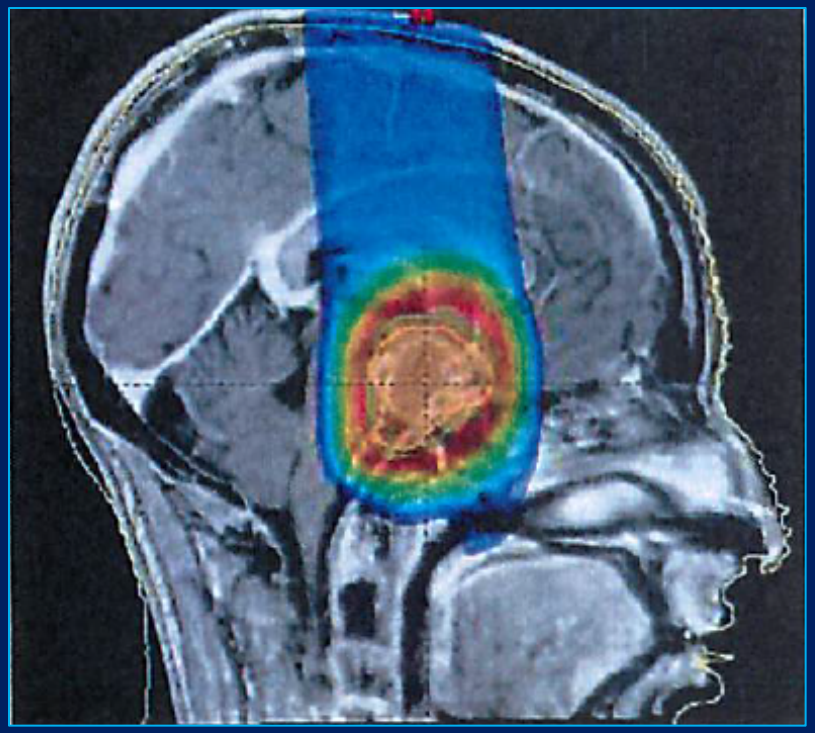
- May 17 2014 – July 9 2014
- 44 days of treatment
  - 54 CGE (Gy) at 1.8 CGE per daily fraction
  - Used a three field 3D conformal proton plan

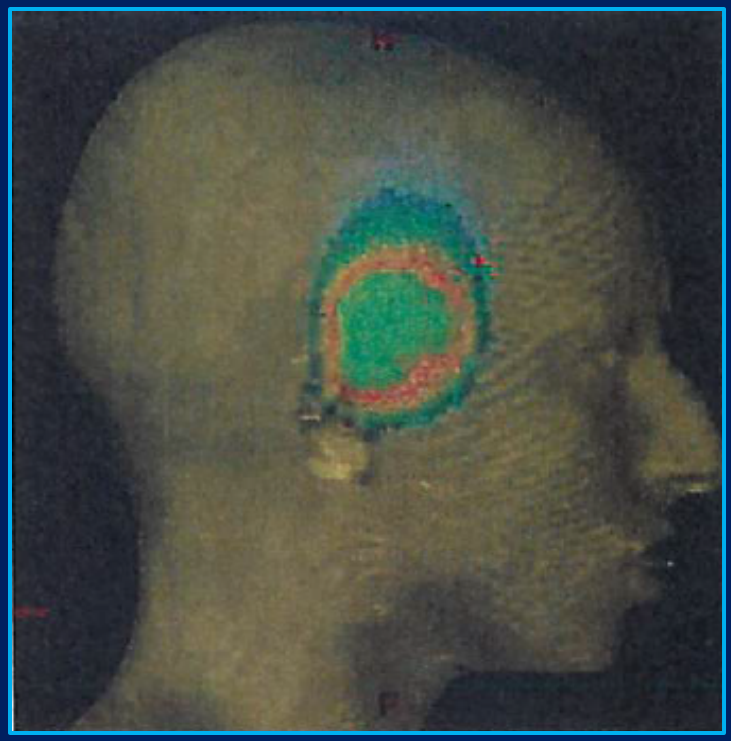
Proton Beams





Proton beams





Post radiation patches (both sides)

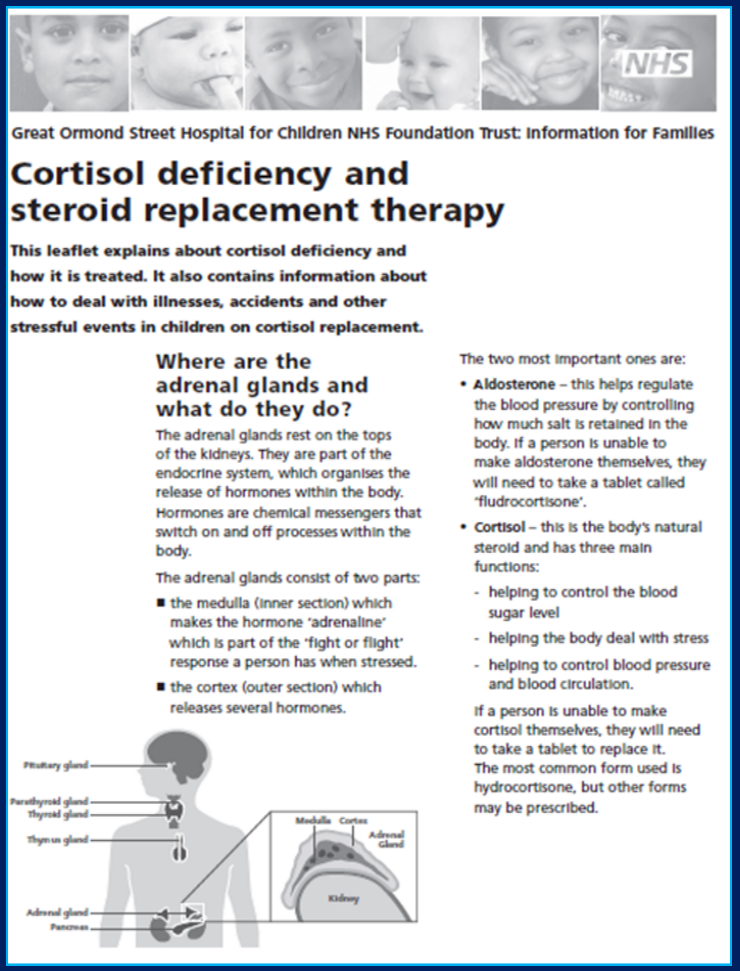
Recommendations after Proton Therapy

- Neurocognitive testing
  - Post treatment baseline
  - Every 2/3 years minimum
- Neuroendocrine testing within 6 months
  - Post treatment baseline
  - Continue every 6/12 at least through puberty
    - Ideally lifelong
      - Pay particular attention to GH
- Ophthalmology and visual acuity

Endocrine follow up

- Back in the UK – July 2014
- Clinical Nurse Specialist review
  - Growth hormone
  - Thyroxine
  - Hydrocortisone
    - Intensive emergency management teaching
      - Medic Alert bracelet
      - Ongoing with school
      - Translations into Spanish

Emergency management



Great Ormond Street Hospital for Children NHS Foundation Trust: Information for Families

Cortisol deficiency and steroid replacement therapy

This leaflet explains about cortisol deficiency and how it is treated. It also contains information about how to deal with illnesses, accidents and other stressful events in children on cortisol replacement.

Where are the adrenal glands and what do they do?

The adrenal glands rest on the tops of the kidneys. They are part of the endocrine system, which organises the release of hormones within the body. Hormones are chemical messengers that switch on and off processes within the body.

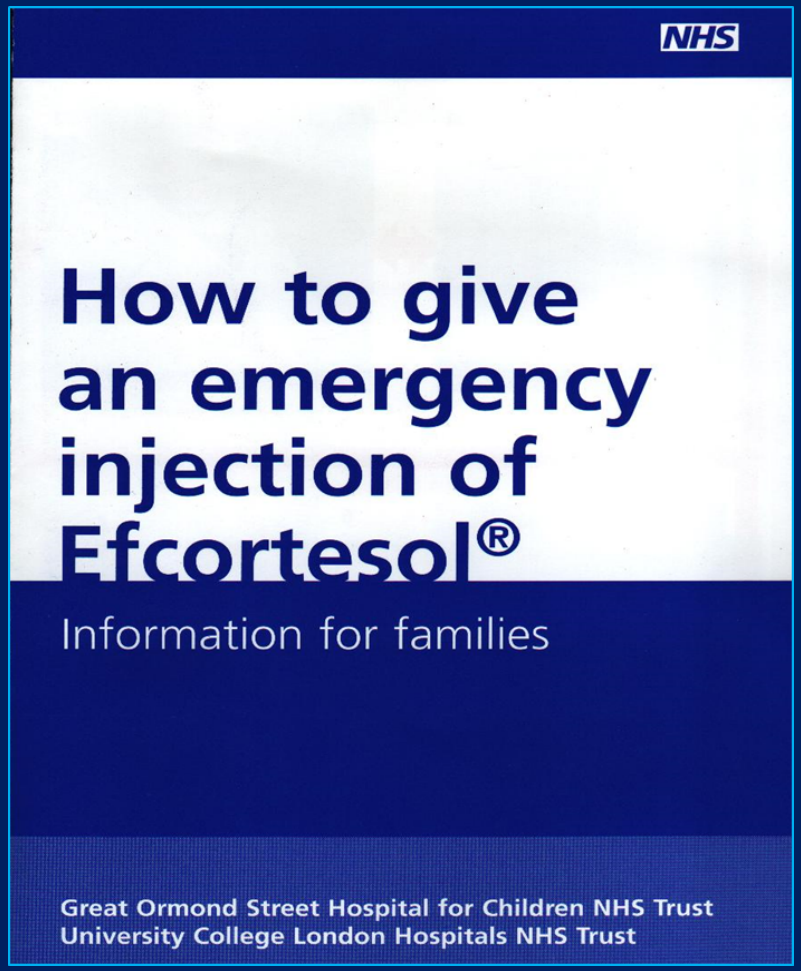
The adrenal glands consist of two parts:

- the medulla (inner section) which makes the hormone 'adrenaline' which is part of the 'fight or flight' response a person has when stressed.
- the cortex (outer section) which releases several hormones.

The two most important ones are:

- **Aldosterone** – this helps regulate the blood pressure by controlling how much salt is retained in the body. If a person is unable to make aldosterone themselves, they will need to take a tablet called 'fludrocortisone'.
- **Cortisol** – this is the body's natural steroid and has three main functions:
  - helping to control the blood sugar level
  - helping the body deal with stress
  - helping to control blood pressure and blood circulation.

If a person is unable to make cortisol themselves, they will need to take a tablet to replace it. The most common form used is hydrocortisone, but other forms may be prescribed.



Emergency services

London Ambulance Service NHS Trust

Patient Specific Protocol
PSP Paediatric Steroid Dependent Crisis

PSP

This protocol has been specifically prepared for STEROID DEPENDENT CRISIS patients and details the treatment to be given in specified circumstances.

Patient's Name: **Date of Birth:**

NHS Number:

Address:

School:-

Local hospital:

Reason for protocol: Administration of IM hydrocortisone in possible adrenal crisis

Specific Treatment / Instructions: Patient may have an adrenal crisis if IM hydrocortisone not administered in an emergency situation

In the event that this child is involved in an accident or develops diarrhoea or vomiting and presents with any symptoms of a steroid dependent crisis whilst at **Home or at School** they are to be administered IM hydrocortisone as detailed over leaf.

Note:- The IM hydrocortisone (Efcortesol) is kept both by the parents and by the school in an emergency pack.

Please transport this child to the above local hospital if possible, otherwise to the nearest paediatric A&E unit.

All other aspects of clinical care remain unchanged.

For further advice if necessary please contact the Endocrine Registrar on call via switchboard at Great Ormond Street Hospital on 020 7405 9200

1. Efcortesol 1ml ampoule (Hydrocortisone 100mg/ml - as sodium phosphate)

Dose: Age 0-1 years 25 mg IM
Age 1-5 years 50mg IM
Age 5+ years 100mg IM

2. Please also administer Glucogel (Hypostop) 25 gram tube, required dose in an emergency - up to 1/3 tube if not already previously administered by carers.

Following administration of the hydrocortisone remove to hospital with full monitoring and oxygen therapy as required.

All other aspects of clinical care remain unchanged.

If required contact EOC and ask for the Clinical Support Desk

PTO for further general info on Steroid Dependent Crisis

London Ambulance Service NHS Trust

Patient Specific Protocol
PSP Paediatric Steroid Dependent Crisis

The symptoms of a Steroid Dependent Crisis

- Weakness
- Mental confusion
- Drowsiness, in advanced cases slipping towards a coma
- Dizziness
- Nausea and/or vomiting
- Headache
- Abnormal heart rate – either too fast or too slow
- Abnormally low blood pressure
- Possibly a fever
- Abdominal tenderness

The causes of a Steroid Dependent Crisis

- Physical shock, e.g. a car accident
- Infection, e.g. flu with a high temperature
- Dehydration, e.g. stomach bug with vomiting

All other aspects of clinical care remain unchanged.

NOT SUITABLE FOR LAS CLINICAL TELEPHONE ADVICE

If required contact EOC and ask for the Clinical Support Desk

Fionna Moore FRCS, FCEM, FIMC RCS Ed
Medical Director
London Ambulance Service NHS Trust

Issue Date:

Emergency hospital letter

Great Ormond Street Hospital for Children NHS Trust

Great Ormond Street
London WC1N 3JH

Tel: 020 7405 9200
Gastroenterology, Endocrinology, Metabolic & Adolescent Medicine (GEMA)
Direct Line: 0207-813-8214

Re:
Diagnosis:
Medications: Hydrocortisone (oral)
Fludrocortisone (oral)
NaCl supplements 5mmol/ml 30% solution:

Instructions for Hospital Doctor

In view of this patients cortisol deficiency, if this patient is brought to hospital as an emergency, the following management is advised:

- If patient drowsy and unresponsive give IM hydrocortisone in the following doses immediately (0-1yr – 25mgs; 1-5 yrs – 50mgs; > 5yrs – 100mgs) if patient has not already had IM hydrocortisone administered by ambulance crew or parents.
- Take blood for U&Es, glucose and osmolality
- If blood glucose is < 2.5 mmol, give bolus of 2mg/kg 10% dextrose
- If patient is drowsy, hypotensive and peripherally shut down, give 20ml/kg of normal saline, insert an IV cannula and then continue with usual dextrose saline infusion
- Continue with bolus IV hydrocortisone at 2mg/kg every 4 hours until patient is tolerating oral fluids and then swap to double usual oral Hydrocortisone doses until patient fully recovered and back to normal self (usually 2-3 days on double usual hydrocortisone doses).
- **Important:** Please admit for a minimum of 12 hours

If there is any doubt about this patients management, advice can be obtained via Great Ormond Street Hospital switchboard (0207 405 9200, asking for the Endocrine Registrar on Call).

Steroid card

Instructions for Hospital Doctor

Dear Doctor,

If this patient is brought to hospital as an emergency the following management is advised:

1) Insert an IV cannula
2) Take blood for U&Es, glucose, and perform any other appropriate tests (e.g. urine culture)
3) Check capillary blood glucose level
4) Give 100 mg hydrocortisone intravenously as bolus (unnecessary if patient has already been given IM hydrocortisone)
5) Commence IV infusion of 0.45% sodium chloride and 5% glucose at maintenance rate (extra if patient is dehydrated). Add potassium depending on electrolyte
6) Commence hydrocortisone infusion (50 mg hydrocortisone in 50ml 0.9% sodium chloride via syringe pump)
7) Monitor for at least twelve hours before discharge

IMPORTANT! If blood glucose is < 2.5 mmol/l, give bolus of 2 ml/kg of 10% glucose

If patient is drowsy, hypotensive and peripherally shut down with poor capillary return give 20ml/kg of 0.9% sodium chloride stat.

If in any doubt about this patient's management, please contact the urgent advice numbers

Useful Contact Numbers:

GOSH Switchboard
Tel: 020 7405 9200

For Urgent Advice:
Tel: 020 7405 9200 and ask to be put through to the endocrine registrar on call

University College Hospital Switchboard
Tel: 0845 155 5000

For Urgent Advice:
Tel: 0845 155 5000 and ask to be put through to the endocrine registrar on call.

Great Ormond Street Hospital for Children NHS Trust and University College London Hospitals NHS Foundation Trust NHS

CORTISOL DEFICENCY

THE OWNER OF THIS CARD IS ON CORTISOL REPLACEMENT THERAPY

Name _____

_____

Address _____

_____

_____

_____

Tel _____

Mobile: _____

Date of Birth ____/____/____

Hospital No _____

Affix photo here

Consultant _____

Hospital _____

Address _____

Tel _____ Fax _____

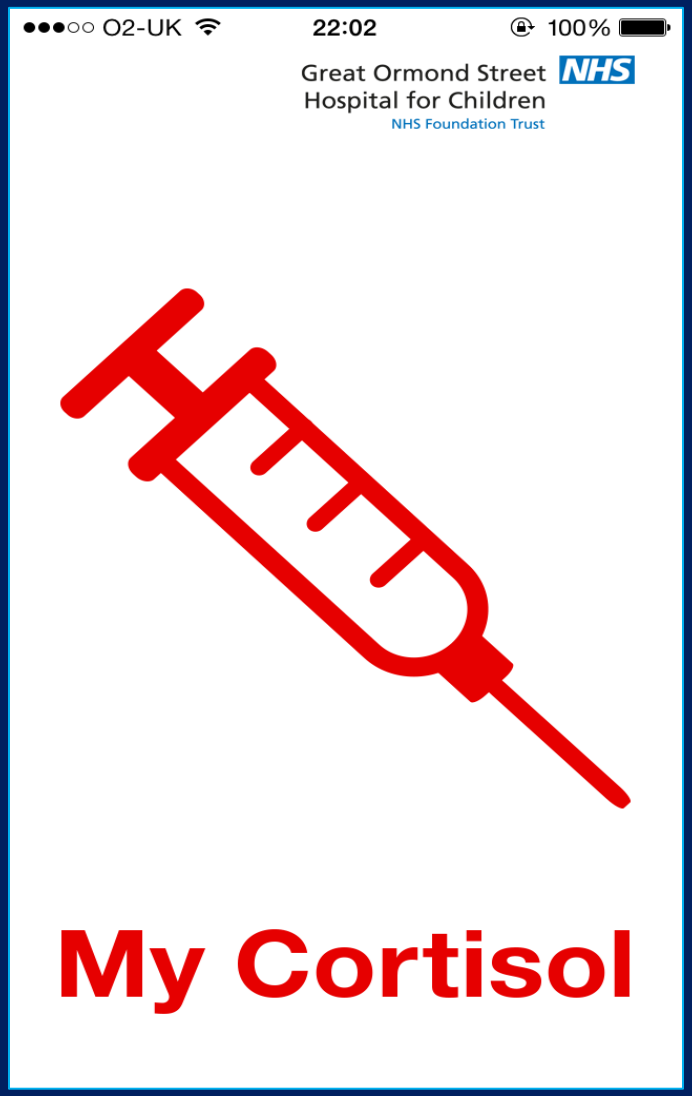
General Practitioner _____

Address _____

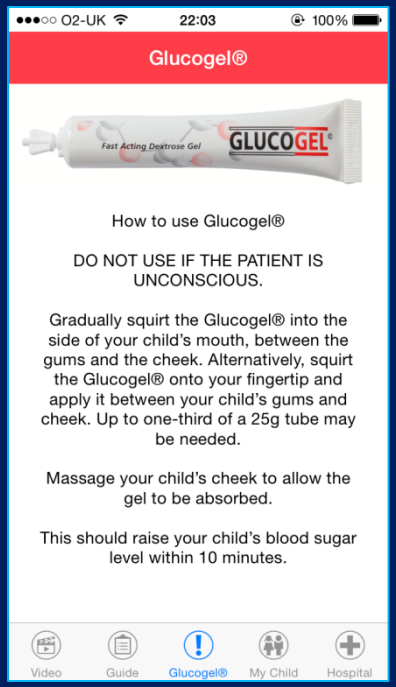
Tel _____ Fax _____

Great Ormond Street Hospital for Children NHS Trust and University College London Hospitals NHS Foundation Trust NHS

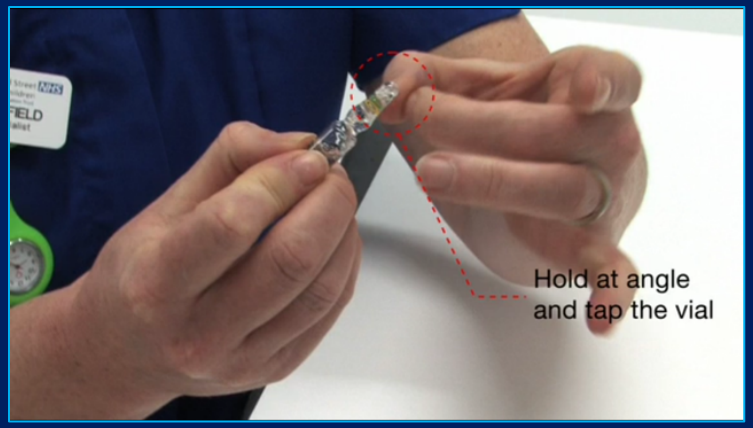
My Cortisol App




O2-UK 22:02 100%
How To - Guide
How to give an emergency Efcortesol® injection
1
Syringe
Needle
Efcortesol
Equipment
2
Tap
Break
Open the vial
3
Attach needle & syringe
4
Draw up
5
6
0.25ml
0.5ml
1ml
Video
Guide
Glucogel®
My Child
Hospital


O2-UK 22:03 100%
Glucogel®
Fast Acting Dextrose Gel
GLUCOGEL
How to use Glucogel®
DO NOT USE IF THE PATIENT IS UNCONSCIOUS.
Gradually squirt the Glucogel® into the side of your child's mouth, between the gums and the cheek. Alternatively, squirt the Glucogel® onto your fingertip and apply it between your child's gums and cheek. Up to one-third of a 25g tube may be needed.
Massage your child's cheek to allow the gel to be absorbed.
This should raise your child's blood sugar level within 10 minutes.
Video
Guide
Glucogel®
My Child
Hospital


O2-UK 22:02 100%
How To - Video
TOUCH TO PLAY VIDEO
Once you have given your child the medication please tap below to call 999 and state your child is having an ADRENAL CRISIS
CALL 999
Video
Guide
Glucogel®
My Child
Hospital


Hold at angle and tap the vial


Attach needle and syringe
GLUCOGEL

Late Effects follow up

after cure - factsheet

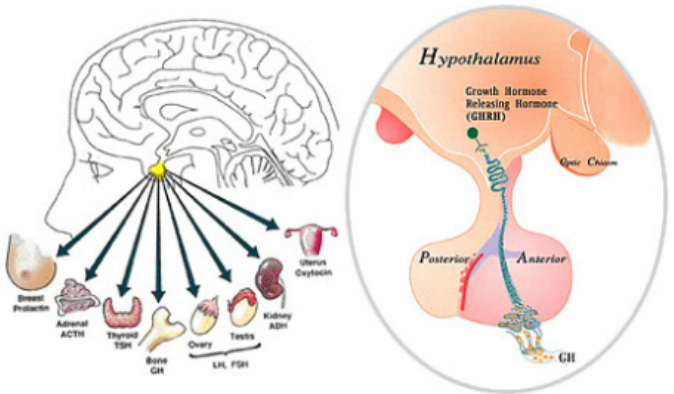
Endocrine: Effects of illness & treatment on growth & development

Treatment for a brain tumour or a tumour in the head and neck area is likely to include radiotherapy to the brain. Treatment may also include chemotherapy (drug treatment) and surgery. All these treatments can affect your hormone levels. The position of the tumour is also an important factor in determining which hormone problems might occur.

Hormones – what are they?

Hormones are chemical messengers produced naturally by the body; they are essential to help you grow, regulate your metabolism, control puberty, fertility and many other things. They also have life saving functions e.g. stress and thirst control. They work by sending specific messages to other organs/parts of your body, in a type of "postal messaging" system.

The pituitary gland and the hypothalmus:



Any illness can cause a temporary "shut down" to the system and alter hormone levels. However after treatment for a brain tumour, and after any treatment where the pituitary has been affected, this can be more permanent.

Luke today

- Continue neuropsyschological and ophthalmological assessments
  - Good cognitive assessments
    - A* at GCSEs
    - A Levels in Barcelona
    - University to study mechanical engineering
- GH discontinued February 2015
- No gonadotrophin deficiency, but Testosterone low normal level
  - USA: 320ng/dl (238 – 850)
- Hydrocortisone increased 7.5 / 5 / 5mg
- Family
  - Increased support wanted - Barcelona

Continuing plan

- Stay off growth hormone
  - ? Start adult GH
- Encourage Thyroxine compliance
- Observe for evolving gonadotrophin deficiency
- Continue annual 24 hour cortisol profiles
  - ? Increase hydrocortisone to 10 / 5/ 5
- Transition – now nearly 19years old

Luke today

Any questions?